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QUAGMIRE OF CONGENITAL CARDIAC DEFECTS IN ASSOCIATION WITH DEXTROCARDIA AND SITUS INVERSUS: RARE CASE REPORT AND REVIEW OF LITERATURE

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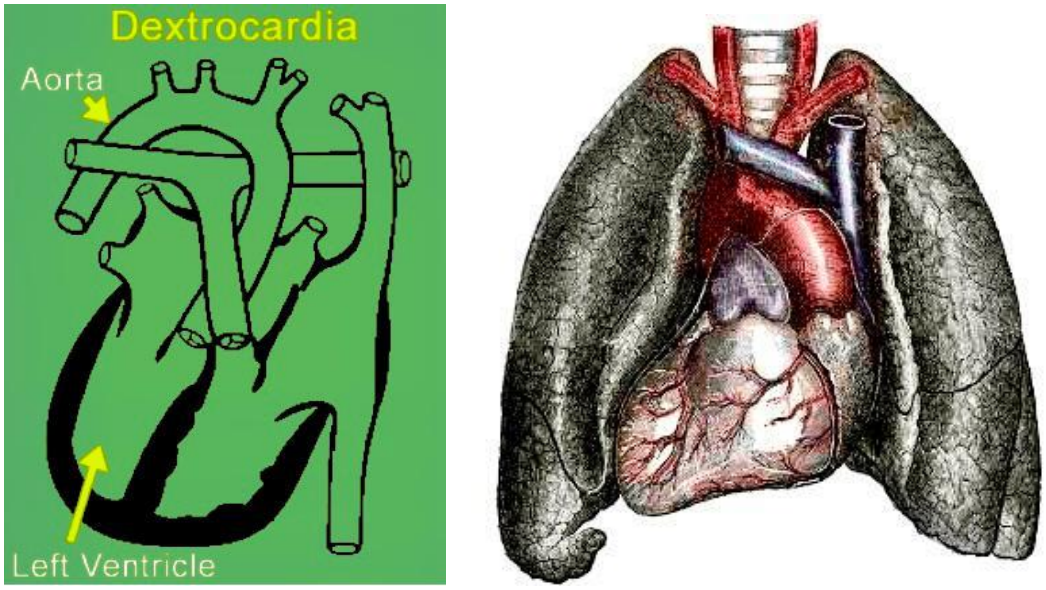
ABSTRACT
Situs inversus (SI) with dextrocardia is the complete reversal of the position of the abdominal and thoracic viscera. It may remain asymptomatic or could be detected early in infancy or childhood, when associated with other congenital malformations especially of the cardiovascular system. Situs inversus with dextrocardia usually exists without co-existing congenital heart disease. We are reporting a case of a nine year female child who presented with complaints of recurrent cough and bluish discoloration of the lips. Examination revealed central cyanosis with grade 3 digital clubbing and a right-sided apical impulse. Chest radiograph showed a right sided heart and Transthoracic Echocardiography (TTE) confirmed multiple cardiac anomalies: Dextrocardia, Situs inversus, Complete ario-ventricular canal defect (CAVCD), Double outlet right ventricle (DORV) with Pulmonary Stenosis (PS).

KEYWORDS: Dextrocardia, Situs inversus, Double outlet right ventricle, Pulmonary valvular stenosis, Complete atrio-ventricular canal defect, CAVCD, Hypoplastic mitral valve annulus.

INTRODUCTION

Dextrocardia is a congenital anomaly in which the heart is situated on the right side of the body[1] (Figure 1).



(A) **(B)**

Figure 1: (A) & (B) Diagrammatic Illustration of Dextrocardia.

SI is a condition in which the major visceral organs are mirrored from their normal positions, that is the morphological left atrium is on the right while the morphological right atrium, the liver and gall bladder are on the left.[2] This is otherwise known as situs inversus totalis (SIT) or mirror image dextrocardia (Figure 2), which is a rare condition with a prevalence of 1:10,000 in some populations.[3, 4] It is generally asymptomatic except when associated with congenital heart defects which is seen in 5 to 10% of cases.[3, 4]

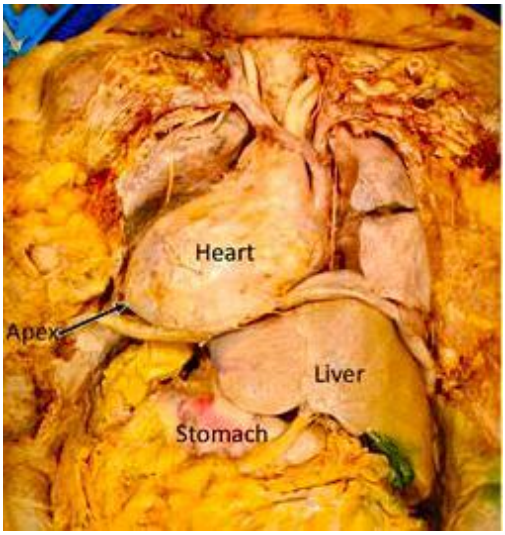


Figure 2: Cadaveric specimen of Dextrocardia with Situs Inversus: The apex of the heart is pointing to the right (Dextrocardia) in the thoracic cavity. The liver is located on the left side and stomach is on the right side.

Classification of congenital dextrocardia[5]

Type I dextrocardia (mirror-image dextrocardia): In Type I dextrocardia, the anatomic right atrium and right ventricle are situated to the left of an anterior to the corresponding systemic chambers. A mirror-image arrangement of the cardiac chambers is present since the frontal relationship of the chambers is reversed, yet, the anteroposterior arrangement is normal.

Type II dextrocardia (dextroversion complex): In Type II dextrocardia, the relations of the cardiac chambers are normal; the right atrium and right ventricle are situated to the right of and posterior to the corresponding systemic chambers. In complete dextroversion the cardiac apex is situated anteriorly and to the right; in incomplete dextroversion or mesoversion, it is located in the substernal region and the longitudinal axis of the heart is parallel to the midsagittal axis of the chest.

Type III dextrocardia (mixed dextrocardia): In Type III dextrocardia is characterized by inversion of the atria alone or of the ventricles alone. The arrangement of the cardiac chambers is therefore in part similar to that of Type I and of Type II dextrocardia.

Type IV dextrocardia (congenital dextroposition): In Type IV dextrocardia, the heart is in the mid chest, but the arrangement of the chambers is normal and the cardiac apex is still directed to the left and anteriorly.

Type V dextrocardia (congenital extrinsic): In Type V dextrocardia, the abnormal position of the heart is due to its displacement by congenital anomalies of the lungs, diaphragm, or chest cage. The rightward displacement may be in the frontal plane only (simple dextroposition) or may occur in both the frontal and horizontal planes (dextroposition with pivotal rotation).

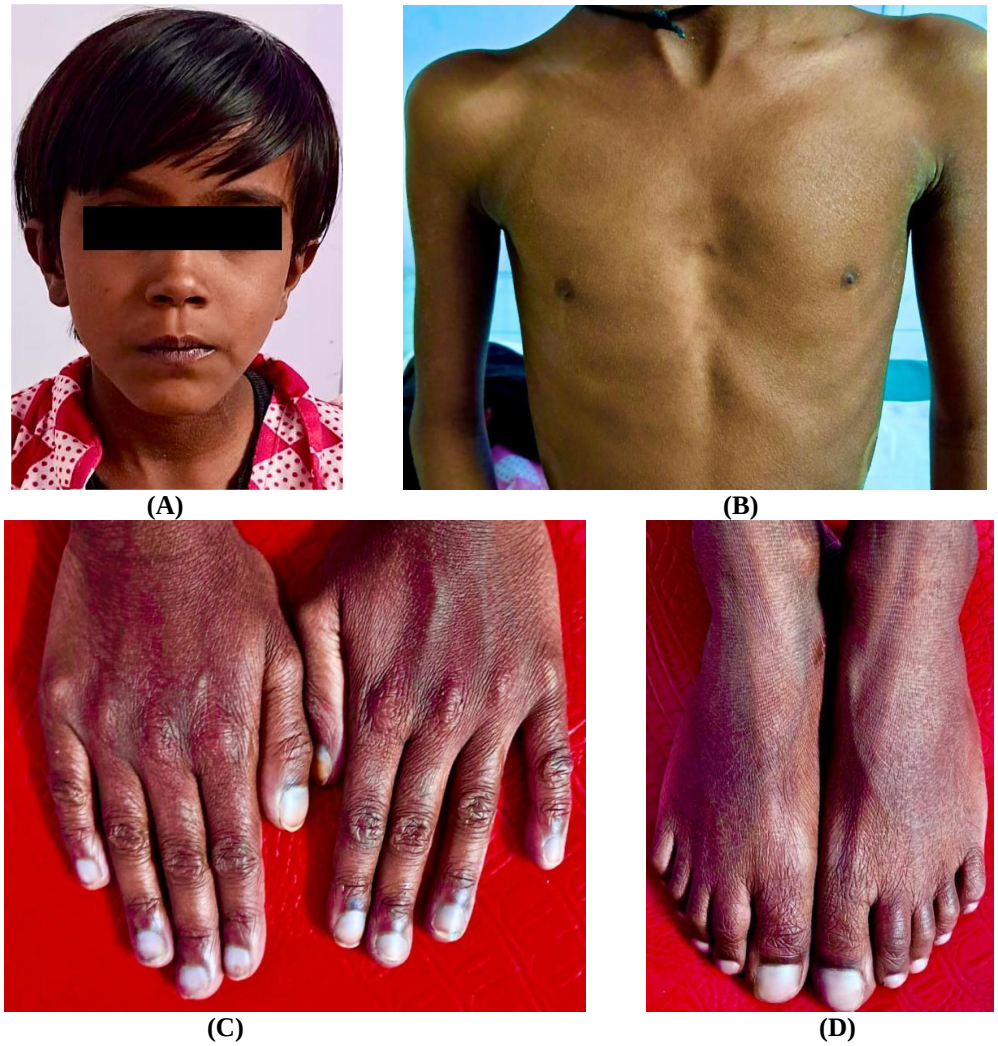
The incidence of cardiac anomalies in the intrinsic group was 79 per cent, and in the extrinsic group 24 per cent.[5] These anomalies were, as a rule, multiple and severe and together formed specific anatomic syndromes usually characterized clinically by dextrocardia, cyanosis, and diminished pulmonary blood flow.[5] The most common entities encountered were tetralogy, single ventricle with pulmonary stenosis, and tricuspid atresia or stenosis.[5]

Here we are reporting a case of a nine year old female child presenting with dextrocardia, SI accompanied with conundrum of congenital heart anomalies.

CASE REPORT

A nine year old female child was referred to us for comprehensive color echocardiography and opinion regarding management of cyanotic congenital heart disease.

The parents provided the history of cyanosis since birth, shortness of breath and failure to thrive. On clinical examination, the child was thin built, very active and lively. Pectus excavatum was noticed alongwith prominent chest wall. Central cyanosis was obvious over her lips, finger tips and toe tips. Conspicuous clubbing was also noticed over nails of fingers and toes (Figure 3).



(A) **(B)**

(C) **(D)**

Figure 3: Facial appearance and other clinical features in our index patient. (A) Facial appearance; (B) Chest wall abnormalities- Pectus excavatum was observed alongwith prominent chest wall; left > right; (C) Clubbing and cyanosis was detected in all the fingers; (D) Clubbing and cyanosis was noticed in all the toes.

The child's weight was 13 kg, height was 168 cm, BP was 90/60 mmHg, HR was 105/min, respiratory rate was 16/min and SPO2 was 65% at room air. All the peripheral pulses were normally palpable without any radio-femoral delay. Cardiovascular examination revealed apical impulse on the right side of chest, in the 5th intercostal space, just medical to the mid-clavicular line. A grade 3/6 ejection systolic murmur was best heard in the right IInd intercostal space. P_2 component was soft. No clicks or gallop sound were heard.

Xray chest (PA) view demonstrated dextrocardia with reduced pulmonary blood flow (Figure 4).

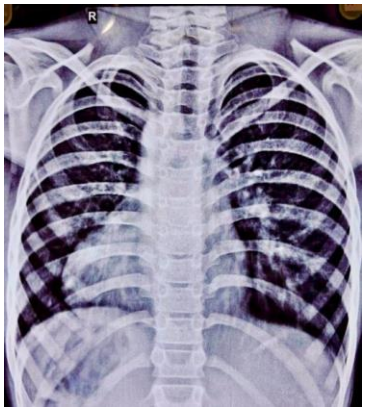


Figure 4: X-ray chest (PA) view- Dextrocardia is recognized. The pulmonary blood flow is decreased.

Resting ECG showed the following features (Figure 5):

- Normal sinus rhythm with a ventricular rate of 96/ min,
- T wave inversion in L1, AVL, V2- V6,
- Tall R waves in V1 (consistent with right ventricular hypertrophy),
- Lack of normal R wave progression in precordial leads (compatible with dextrocardia).

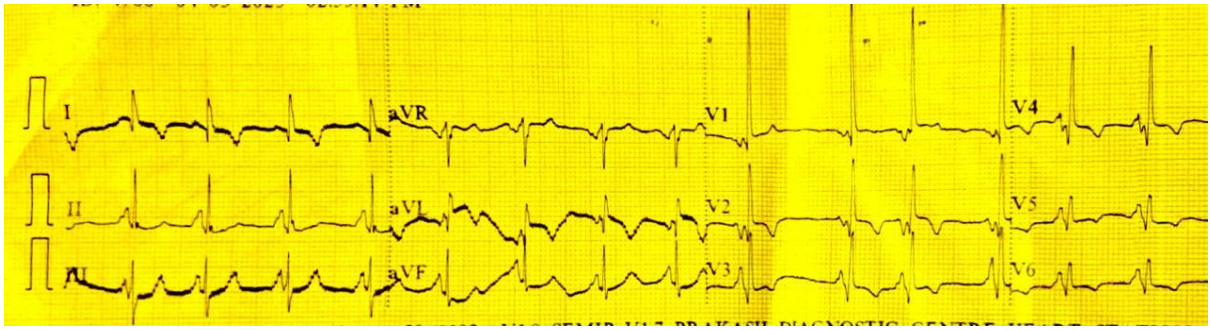


Figure 5: Resting ECG demonstrated: Normal sinus rhythm with a ventricular rate of 96/ min, T wave inversion in L1, AVL, V2- V6 , Tall R waves in V1 (consistent with right ventricular hypertrophy), lack of normal R wave progression in precordial leads (compatible with dextrocardia).

Transthoracic Echocardiography

All echocardiography evaluations were performed by the author, using My Lab X7 4D XStrain echocardiography machine, Esaote, Italy. The images were acquired using a adult probe equipped with harmonic variable frequency electronic single crystal array transducer while the subject was lying in supine and left lateral decubitus positions.

Conventional M-mode, two-dimensional, pulse wave doppler (PWD), continuous wave doppler (CWD) and sequential segmental transthoracic echocardiography was performed in the classical subcostal, parasternal long axis (LX), parasternal short axis (SX), 4-Chamber (4CH), 5-Chamber (5CH) and suprasternal views.

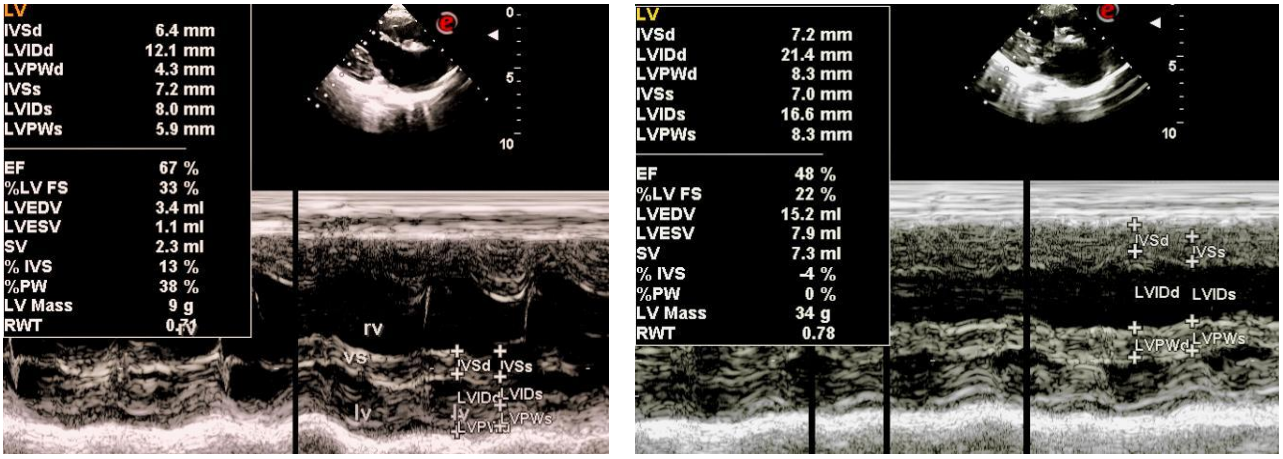
M-mode Echocardiography

M-mode echocardiography of left and right ventricle was achieved for estimating various ventricular parameters (Table 1, Figure 6).

Table 1: Calculations of M-mode echocardiography.

Measurements	**LV**	**RV**
IVS d	6.4 mm	7.2 mm
ID d	12.1 mm	21.4 mm
PW d	4.3 mm	8.3 mm
IVS s	7.2 mm	7.0 mm
ID s	8.0 mm	16.6 mm
PW s	5.9 mm	8.3 mm
EF	67 %	48 %
%FS	33 %	22 %
EDV	3.4 ml	15.2 ml
ESV	1.1 ml	7.9 ml
SV	2.3 ml	7.3 ml
Mass	9 g	34 g

IVS, interventricular septum, ID, internal dimension; PW, posterior wall, d, diastole; s, systole; FS, fractional shortening; EDV, end-diastolic volume; ESV, end systolic volume; SV, stroke volume; EF, ejection fraction.



(A) **(B)**

Figure 6: M-mode Echocardiography. (A) LV volumetric estimation; (B) RV volumetric estimation.

Summary of M-mode echocardiography

M-mode echocardiography depicted dilated RV with mildly reduced RVEF (48 %). On the contrary the LV was hypoplastic with normal LV systolic function LVEF (67 %).

2Dimensional-Transthoracic Echocardiography

2Dimensional transthoracic echocardiography (TTE) was conducted in explicit detail and it demonstrated the following features (Figures 7-18):

- Dextrocardia
- Situs inversus
- Concordant L – Bulboventricular Loop,

AV concordance

VA discordance

- L-loop ventricles
- Morphological RV is to the left of morphological LV - Confluent pulmonary arteries
- Double outlet right ventricle (DORV) - Both the great arteries are arising from the morphological right ventricle.
- A subaortic conus was recognised
- Malposition of great arteries

Aorta is anterior and to the right

Pulmonary artery is posterior and to the left

- Complete atrio - ventricular canal defect (CAVCD) - Rasteilli Type B

The chordae of left AV valve (Tricuspid valve) were attached to the lateral wall of RV and RV apex. MV chordae were attached to ventricular septal crest and lateral wall of LV.

- Common AV valve (CAAV) orifice
- Common AV valve with separate MV (Right AV valve) and TV (Left AV valve) orifices
- MV orifice was hypoplastic and TV leaflets were large and thickened.
- Atrial septal defect (large)

Size: 9.8 mm.

Ostium primum type.

Lt. to Rt. shunt.

- Ventricular septal defect (large)

Size 15.9 mm.

Inlet type

Lt. to Rt. shunt.

- Left AV valve (TV) regurgitation (mild)

On color flow mapping a mild, central tricuspid regurgitation jet was identified in RA. The jet area was 2.80 sqcm.

- Pulmonary valvular stenosis (severe)

PV domed

Peak/mean gradient across PV = 83/64 mmHg.

- Hypoplasia of PV annulus, MPA, LPA and RPA

PV annulus (D): 4.0 mm

MPA (D): 5.3 mm

LPA (D) : 3.6 mm

RPA (D): 3.5 mm

- Morphological RV was dilated with mildly reduced - RVEF (48 %)

Morphological LV was hypoplastic

Normal LV systolic function LVEF (67 %)

- Dilated RA, hypoplastic LA, LV.

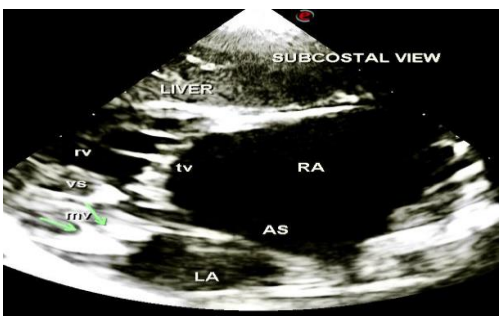


Figure 7: Dextrocardia. Subcostal view identifies dextrocardia.

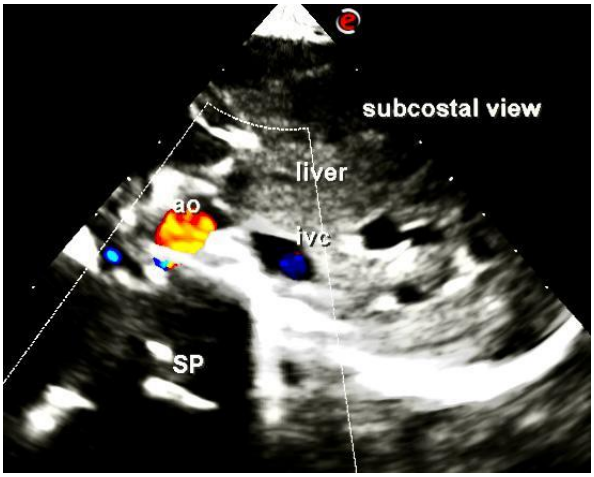
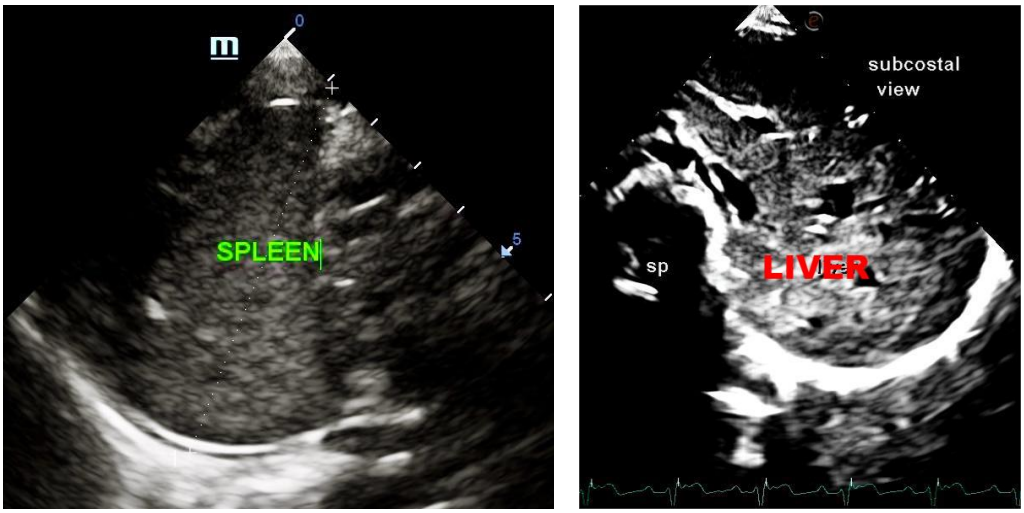


Figure 8: Situs Inversus: In the subcostal view aorta is on the right side and inferior vena cava (ivc) is on the left side. Liver is also left sided; ao, aorta; ivc, inferior vena cava; sp, spine.



(A) (B)

Figure 9: Situs Inversus. Ultrasound of abdomen reveals: (A) Spleen was on the right side; (B) Liver was on the left side; sp, spine.

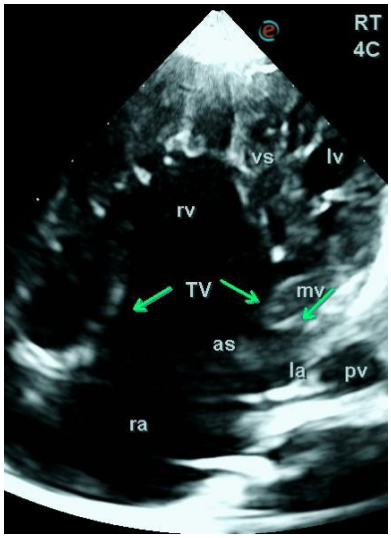
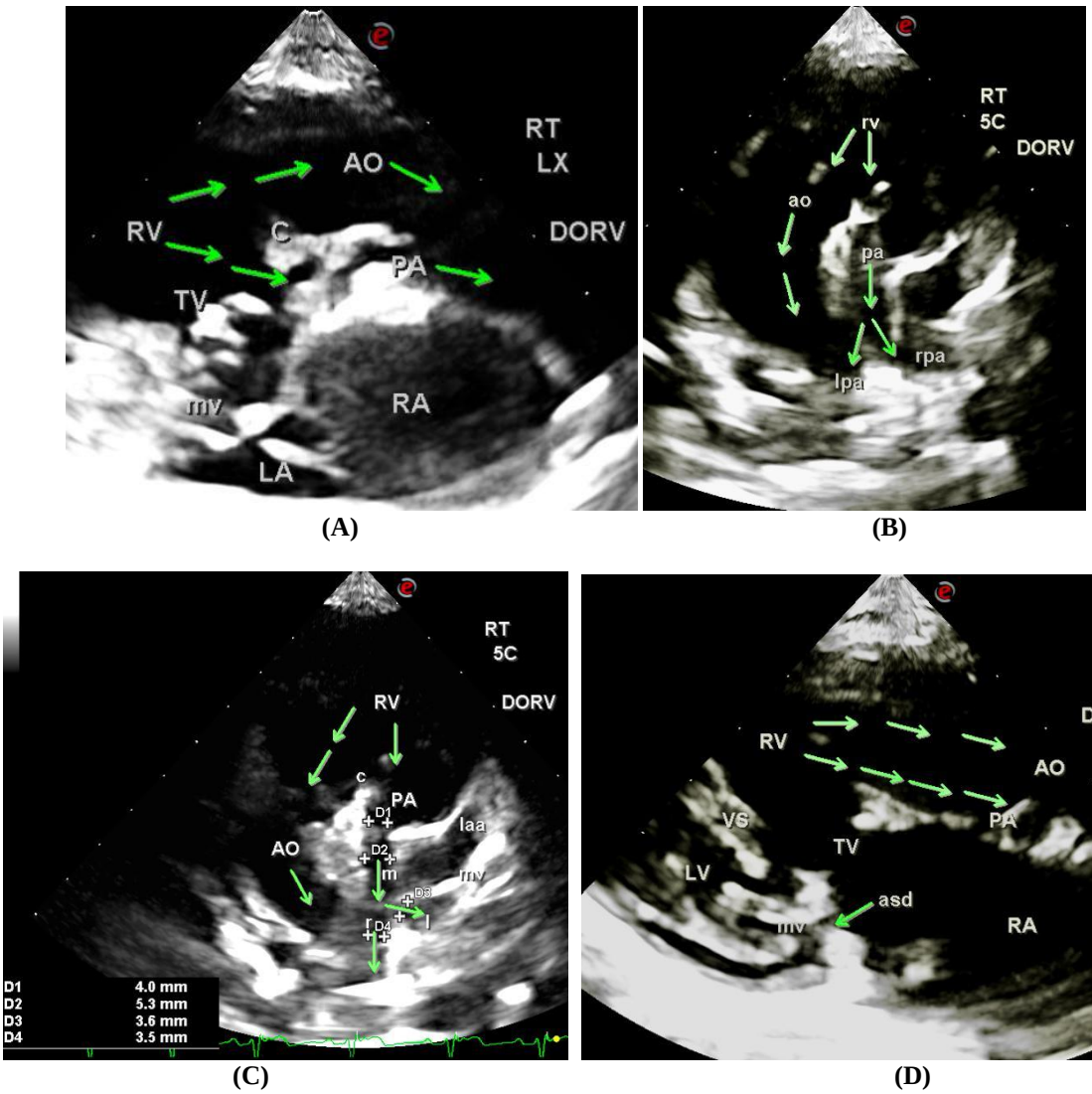


Figure 10: Apical 4C view from right chest with cursor probe placed at 9 o'clock position. Morphological right ventricle was dilated and morphological LV was hypoplastic; la, left atrium; lv, left ventricle; ra, right atrium; rv, right ventricle; vs, ventricular septum; TV, tricuspid valve; as, atrial septum.



(A) **(B)**

(C) **(D)**

Figure 11: Double outlet right ventricle. (A) and (B) In the right LX and right apical 5C views, both great arteries are arising from morphological RV. Aorta was anterior and pulmonary artery was posterior and hypoplastic. Pulmonary valve was domed. A subaortic conus was also recognized. (C) In the right apical 5C view, the diameter of hypoplasic pulmonary arteries was estimated. PV annulus, MPA, LPA and RPA diameter was 4.0 mm, 5.3 mm, 3.5 mm and 3.6 mm respectively; (D) Another image of DORV in the LX view from right chest; DORV, double outlet right ventricle, Ao, aorta, PA, pulmonary artery; C, conus; LA, left atrium; TV, tricuspid valve; RV, right ventricle; mv, mitral valve; asd, atrial septal defect; l, left pulmonary artery, r, right pulmonary artery; vs, ventricular septum.

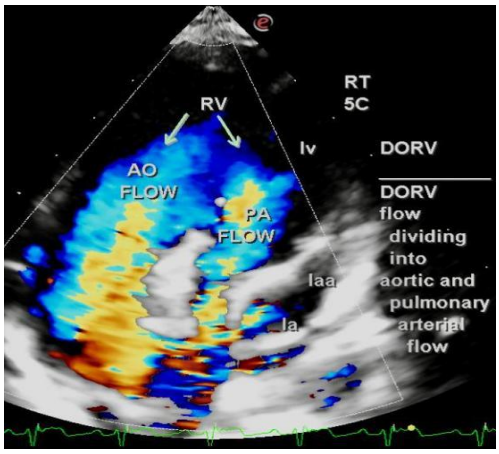


Figure 12: Color flow mapping in apical 5C view from right chest. The classical movement of blood flow in DORV is depicted. The anterior aortic flow is non turbulent and the posterior pulmonary arterial flow is highly turbulent because of severe pulmonary valvular stenosis; la, left atrium; lv ,left ventricle; laa, left atrial appendage; AO, aorta; PA, pulmonary artery.

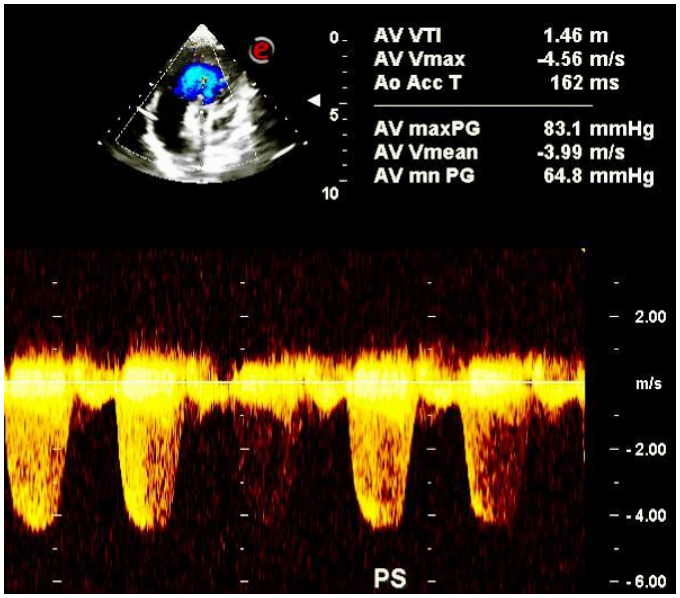
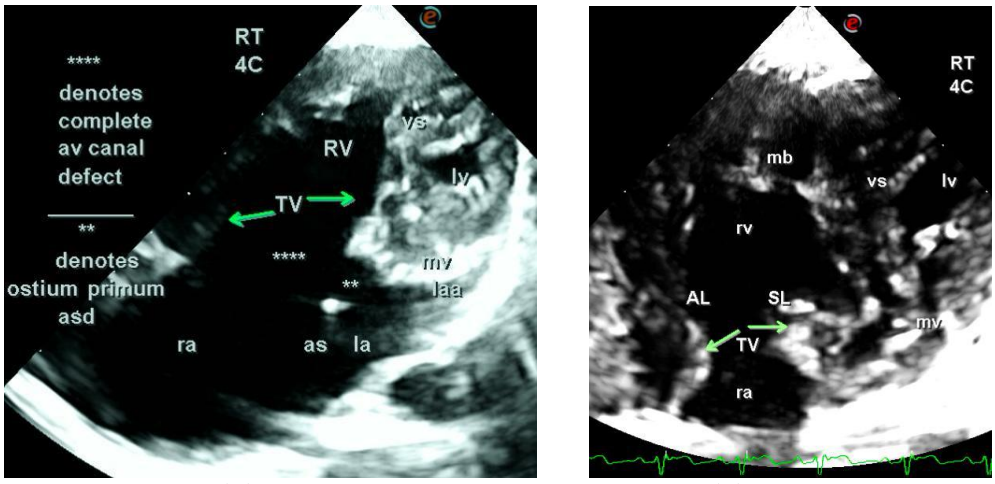


Figure 13: Color flow mapping across pulmonary valve identifies pulmonary valvular stenosis with a peak/mean gradient of 83.1/ 64.8 mmHg; PS, pulmonary stenosis.



(A) **(B)**

Figure 14: Complete atrio-ventricular canal defect (CAVCD). (A) Apical 4C view from right chest discerns the ostium primum ASD (**)and CAVCD (****) ; (B) Apical 4C view from right chest portrays common AV valve with two components: large anterior and septal leaflet of TV and bileaflet MV; mv, mitral valve; lv, left ventricle; vs, ventricular septum; mb, moderator band ; rv, right ventricle; ra, right atrium; AL, anterior leaflet of TV; SL, septal leaflet of TV; as, atrial septum; TV, tricuspid valve.

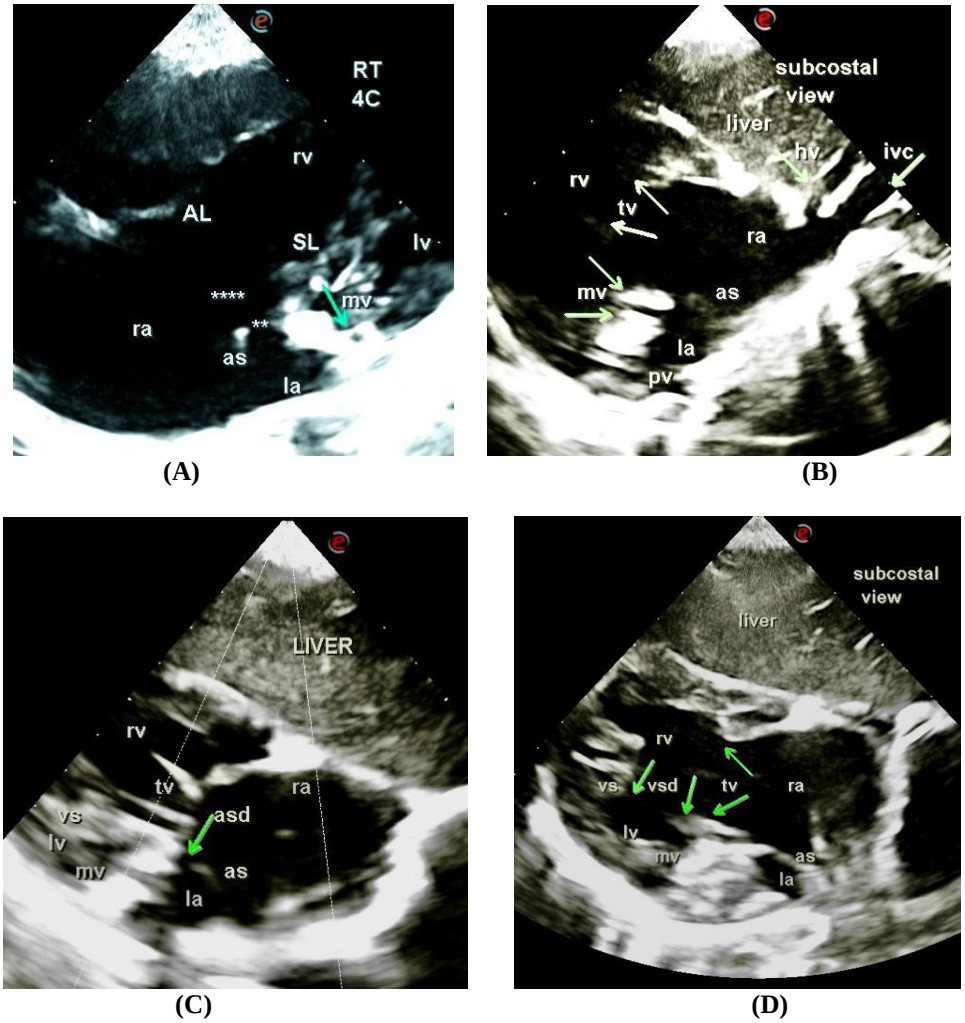


Figure 15: Complete AV canal defect. The apical 4C view from the right chest and subcostal view delineates CAVCD with large ASD and VSD. Dilated RV and small LV. Large TV leaflets and hypoplastic MV annulus was also recognized. AL, anterior leaflet; SL, septal leaflet; **, CAVCD; **, ostium primum ASD; hv, hepatic vein; ivc, inferior vena cava; la, left atrium, lv, left ventricle; ra, right atrium; rv, right ventricle; as, atrial septum; vs, ventricular septum; tv, tricuspid valve; mv, mitral valve.**

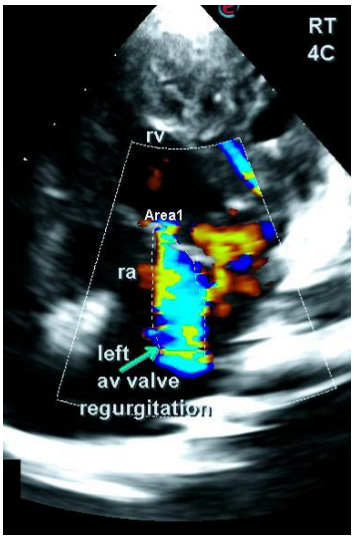


Figure 16: Left AV valve (TV) regurgitation. Apical 4C view from right chest shows mild TV regurgitation.

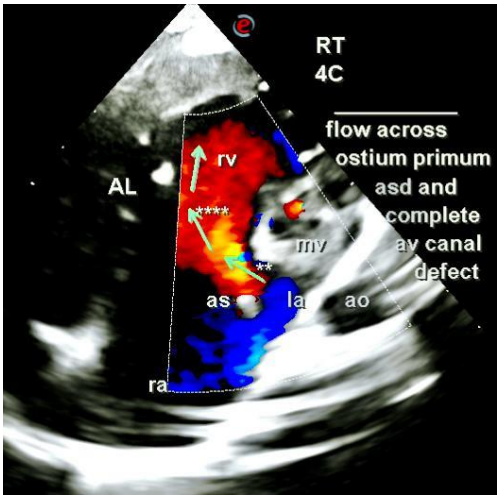
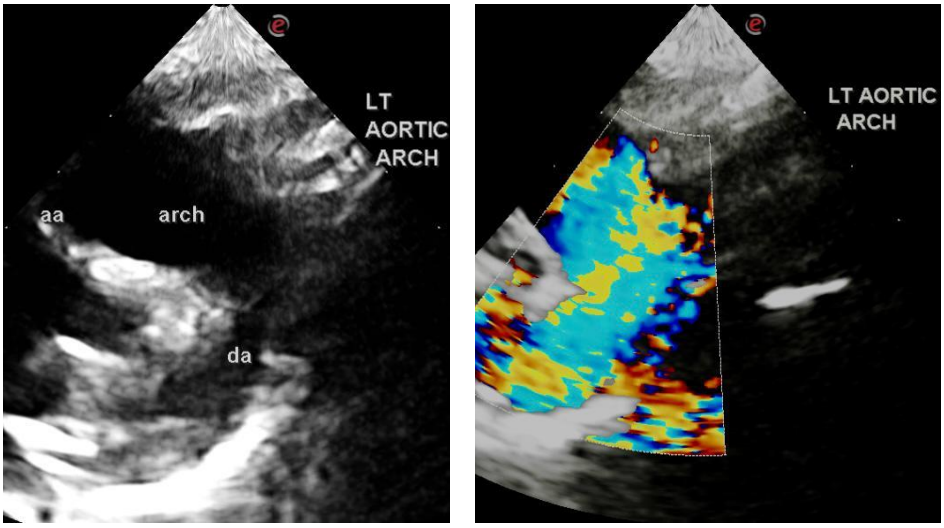


Figure 17: Color flow mapping across ostium primum ASD denoted as (**) and complete atrio-ventricular canal defect (CAVCD) denoted as (****) . Green arrows portray the flow of blood across ASD and CAVCD ; la, left atrium; ra, right atrium; rv, right ventricle; as, atrial septum; ao, aorta; mv, mitral valve; AL, anterior leaflet of tricuspid valve.



(A) **(B)**

Figure 18: Suprasternal view. (A) Left aortic arch; (B) Color flow mapping demonstrating non turbulent blood flow in the left aortic arch; as, ascending aorta; da, descending aorta.

Summary of color echocardiography

On TTE, in our index patient, we detected numerous cardiac defects: Dextrocardia, Situs inversus, Malposition of great arteries, DORV, PS, CAVCD-Rastelli Type B, Hypoplastic MV annulus, LA, and LV accompanied by left aortic arch.

DISCUSSION

SIT is a rare congenital abnormality characterized by a mirror-image transposition of both the abdominal and the thoracic organs. This is a global defect of situs orientation, as the failure to generate normal left-right asymmetry results in a spectrum of laterality disturbances.[6] This condition might cause difficulties during diagnostic and therapeutic procedures.[7]

Nomenclature

The body of vertebrate species shows an external bilateral symmetry. Therefore, it is surprising that humans and all other vertebrates present an internal asymmetry: nearly all visceral organs are left–right (LR) asymmetrical in their anatomy and placement.[6] Any failure in the normal left-right asymmetry stands within a spectrum of laterality disturbances. Heterotaxia is the common term used for these disorders. It encompasses a wide variety of disorders which may be partial or complete. The asymmetry of unpaired organs (heart, liver, spleen, stomach, bowels, aorta, v. cava) seems to be logic and "unavoidable". The asymmetry of paired organs seems less obvious, but they have distinct left and right forms. The lungs have major differences (three lobes vs two lobes), but even kidneys show some asymmetry. Especially the vascular differences between

left and right kidneys have some technical importance, eg, for transplant surgeons.

The complete reversal of normal organ position, including both the thoracic and abdominal organs, is called situs inversus totalis. Other types of situses are situs solitus (normal) and situs ambiguous (indeterminate), which is characterized by isomerism, heterotaxy, and multiple malformations in one or more thoracic or abdominal organs.[8]

Risk factors - Genetic mutations

In a population-based study using multivariate analyses, six risk factors for SIT were identified: family history of heart defects, family history of noncardiac anomalies, maternal diabetes, antitussive use, paternal smoking, and low socioeconomic status.[9] These factors might have some connection with genetic mutations, as major advances in the identification of gene mutations in animals and humans support a growing body of knowledge about the causes of laterality defects.[10]

SI is seldom seen in identical twins[7, 11], even though in conjoined twins, the incidence of SIT is high.[12]

Laterality is established early in development and is orchestrated by a cascade of signal molecules and genes. Importantly, the neurotransmitter serotonin (5HT) also plays a critical role in this signalling cascade that establishes laterality.[13] Sidedness is established at the time of primitive streak formation (gastrulation) and is regulated by genes, such as Nodal and PITX2, which become restricted in their expression (Figure 19).

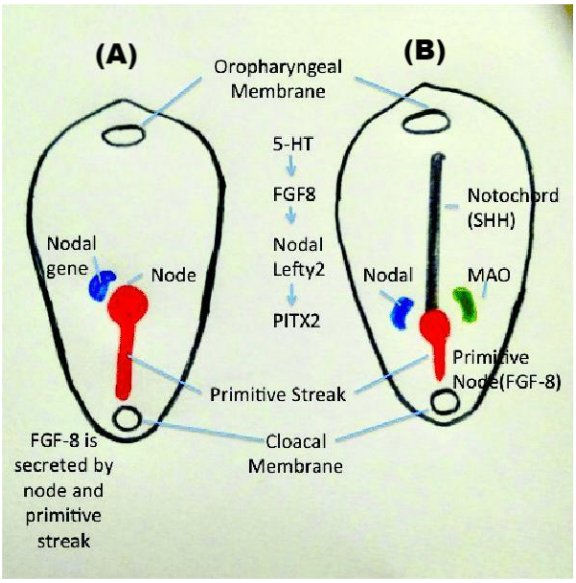


Figure 19: Gene expression patterns for establishing the left-right axis at the 3rd week of fetal development.

Partial reversal of left–right asymmetry is more often associated with other defects than complete asymmetry (SIT).[13] The discovery of these cascades has also shed light on the cause of situs defects in conjoined twins.[14]

Genetic deletions of KIF3-A or KIF3-B, two microtubule-dependent kinesin motor proteins, resulted in a randomization of the situs of the viscera.[15–17] Most cases of heterotaxy are sporadic occurrences without a recognizable cause.[9]

Epidemiology

The incidence of all lateralization defects is approximately 1:15,000 according to the Orphanet.[18] Heterotaxy is present in approximately 1 in 10,000 births.[19] The incidence of situs inversus itself is reported to be 1:6500 to 1:25,000, see Table 2.

Table 2: Estimated Frequency of Left-Right axis Disorders.[20]

Heterotaxy	1:10 000			
Situs ambiguous	1:10 000	1:15 000		
Situs inversus totalis	1:20 000	1:6-8000	1:6500	1:25 000

SIT in inherited in an autosomal recessive pattern[20] and is slightly more frequent in males: 1.5:1.[21]

Classification of situs inversus

Situs inversus could be classified into the following:

- SIT, where the abdominal organs are mirror images of the normal anatomy, as well as dextrocardia.[22]
- SI with levocardia, where the base-to-apex axis points to the left while the abdominal viscera are reversed.[22]

- Situs ambiguous. In these patients, the liver may be midline, asplenia or polysplenia, unclear cardiac morphology, or mal-rotated gut.[22]

Compared to the other types, SIT is the commonest.[23]

Associations

The common congenital cardiac anomalies associated with dextrocardia with situs inversus are atrial situs solitus (93%), discordant AV connection (44%), and discordant Ventriculo-Atrial (VA) connection (30%). Congenitally corrected transposition of great arteries (TGA) occurs in less than 1% of all forms of congenital heart disease.[24] Certain congenital anomalies such as polysplenia (left isomerism)/asplenia (right isomerism) or Kartagener's syndrome often leading to infection of the paranasal sinuses and lungs) are known to occur.[25] About 25% of individuals with situs inversus have an association with primary ciliary dyskinesia. SIT with primary ciliary dyskinesia together known as Kartagener`s syndrome characterized by the triad of situs inversus, chronic sinusitis, and bronchiectasis.[26]

Situs Invesus and Organ Transplantation

Organ transplantation in people with SIT presents unique challenges due to the mirror-image reversal of their organs. Surgeons must carefully assess the changed anatomy using modern imaging investigations and have particular surgical expertise to negotiate the challenges of transplant procedures in these patients. Posttransplant care and surveillance are critical for SIT people, as they may experience a higher risk of complications that need rapid intervention.[27, 28] In circumstances where multi-organ transplants are required, coordinating numerous surgeries adds complexity to the treatment plan.[29, 30] Continued research and developments in transplant medicine are required to improve outcomes for SIT patients through innovative surgical procedures and individualized treatment plans. Addressing these concerns can assist to improve the care and treatment outcomes of SIT patients receiving organ transplants.

CONCLUSION

Dextrocardia with situs inversus is a rare congenital malformation that must be thoroughly evaluated when noticed because it may result in fatal outcomes in rare instances. There is a need for a complete and elaborate diagnostic work up of suspected cases by various imaging modalities, so that the diagnosis cannot be missed. Doctors should encourage routine medical examinations for their patients, which could help identify this anomaly, thereby preventing the wrong diagnosis and possibly death due to delay in management. To confirm the suspected cases of situs inversus, imaging studies should be done, such as a chest x-ray or ultrasound, and a referral may be made to a cardiologist. Imaging studies will also rule out the possibility of random arrangement of the organs, or heterotaxy, which has a much higher risk for serious medical complications. Documenting situs inversus in an individual is essential to avoid any inadvertent clinical or surgical mishap. Our case is one of the rarest reported combinations of Dextrocardia, Situs inversus, Malposition of great arteries, DORV, PS, CAVCD-Rasteilli Type B with Hypoplastic MV annulus and left aortic arch.

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